

Summary

Variant Creutzfeldt-Jakob disease (vCJD) is one of the prion diseases (disorders that arise from an irreversible mutation in the prion protein). The BSE epidemic in the United Kingdom is generally acknowledged to have been the cause of this disorder. Variant CJD displays a different clinical and pathological picture from the classic form of Creutzfeldt-Jakob disease (CJD) in that young people can also contract the disease and in many patients vCJD initially manifests itself in behavioural changes, which result in a visit to a psychiatrist. The patients usually die after a period of just over one year. A decade after it was first reported, vCJD remains a progressive and invariably fatal disease.

The ability to detect abnormal prion protein in lymphoid tissue from vCJD patients by means of the so-called Western Blot test paves the way for laboratory testing. The British government has commissioned retrospective research on tonsil and appendix tissues removed during operations. Extrapolation of the results of this research suggests that abnormal prion proteins are detectable in 237 per million inhabitants of the United Kingdom. No data of this kind are available for the Netherlands. The prevalence here is presumed to be lower than in the United Kingdom, but how much lower is not known.

It is highly probable that transmission of vCJD via blood transfusions has occurred. In the United Kingdom, two recipients of cellular blood products (derived from a donor who was subsequently to develop vCJD) have died from vCJD. The chances of these deaths being unrelated to the receipt of the blood transfusions are extremely slim. In a third recipient, who died of a different cause, abnormal prion proteins were detected in spleen and lymph nodes.

Transmission via blood transfusion has led to calls for a rapid, non-invasive test based on the detection of abnormal prion protein in the blood (the previously mentioned Western Blot test does not have these characteristics). Various companies and university groups are busy developing just such a test and rapid advances have been made. It is expected that a test suitable for use in a blood bank will be on the market within a few years. The test characteristics are not known at present, nor is it clear what the costs of introducing such a test will be. Nevertheless, there will probably be great pressure to introduce this test in the United Kingdom. The case for and against testing will probably also be debated in France and Ireland in the relatively near future. This issue will also need to be considered in the Netherlands, partly because introduction elsewhere may act as a precedent.

The framework for decision-making in this area will be defined by government's constitutional and internationally enshrined responsibility for the availability and safety of blood supplies. That responsibility is fleshed out in the Blood Supply Act (Wibv). Implementation has been entrusted to the Sanquin Blood Supply Foundation. The basic premise of the Blood Supply Act is that the supply of blood in the Netherlands "must satisfy stringent safety and quality requirements". The government has repeatedly emphasised that this does not mean maximum safety, but optimum safety. After all, maximum safety would mean ruling out every possible risk, regardless of the relationship between the health benefit that stands to be achieved and the costs and other disadvantages associated with this measure. The limited financial resources available within the healthcare system are not the only reason why such an approach cannot be justified.

Tests are performed on donors and their blood in order to protect the recipients against blood-borne diseases. In the case of serious, untreatable disorders, however, a dilemma arises. After all, the corollary is then that a positive (i.e. abnormal) test result may have extremely far-reaching consequences for the donor. It is very emotionally distressing to discover that you have an increased risk of developing a serious disease which can neither be treated nor prevented. This information may also have negative implications for the person concerned as far as work and insurability are concerned, or otherwise may lead to exclusion and stigmatisation. This makes testing for such disorders, and therefore also for vCJD, both morally and legally problematic.

Furthermore, if the introduction of a test for vCJD were to result in large numbers of donors being deterred from continuing to give blood, this could jeopardise the maintenance of adequate blood supplies. For various reasons, it is unacceptable – also from a legal standpoint – to test the donor and then not inform him/her of the result. The question therefore arises as to whether it is sensible to test for vCJD if the price to be paid for securing this greater safety would be that insufficient blood is available to meet the needs of patients.

A further significant problem in this connection is the large number of false-positive results that can be anticipated. If prevalence is low, this problem cannot be avoided even by using a relatively specific test. Also to be taken into consideration is the fact that an initial test is probably less discriminatory and that a confirmatory test may not yet be available at first. Besides causing unnecessary anxiety, false-positive results also result in further exclusion of donors. False-negative results, which probably occur far less often, engender unwarranted reassurance in the donor and a lack of certainty in the recipient.

The decision-making over the possible introduction of a test for vCJD will in any case take place under conditions of great scientific uncertainty. Relevant considerations are the question of whether the greater safety for recipients offsets the disadvantages of testing for the donors, the extent to which such a test will, in fact, undermine donor willingness, and the cost-effectiveness of testing for vCJD. Further research – both into attitudes among donors and into the prevalence of abnormal prion proteins in the Netherlands – can only go so far in helping to reduce the continuing uncertainties. The question of what we are to understand by 'optimum' blood safety in

this context is still unresolved. This issue is further complicated by the fact that public perception of risk is also shaped by all manner of affective (and consequently less 'rational') factors.

Under the Blood Supply Act, the government can decide that blood donors must be tested for vCJD. However, the Sanquin Blood Supply Foundation – the privatised blood supplier – also bears a responsibility of its own. Clearly, concerted policy development is desirable, also because of such aspects as financing and liability.

If it is decided to introduce testing, additional measures will be required in order to minimise undesirable consequences for donors and others. First of all, the donor must be adequately informed about the test for vCJD and its possible implications. Then donors with a positive test result must be offered counselling services. Donors who test positive must be assured of adequate care and protected against forms of stigmatisation and social exclusion. We must assess whether (and under what circumstances) it is desirable to trace and inform the recipients of earlier transfusions with blood from donors who have been found to test positive. For the protection of third parties, steps must be taken to prevent further transmission of abnormal prion proteins in the course of delivering medical care to donors who test positive.

If the outcome of the decision-making is that a test for vCJD is not introduced in the Netherlands (for the time being, at least) because such a measure would not be consistent with a policy that is geared to optimum safety, or because the negative consequences of its introduction outweigh the positives, then the emphasis will shift from the consequences for the donor to the implications for the recipient. In the event of adverse health effects that could have been avoided by testing for vCJD, Sanquin (and possibly also the government) may be held liable. The precise legal implications in this scenario require closer consideration. From a moral perspective, consideration also needs to be given to the possibility of compensating people who contract vCJD as a result of the decision not to test donors.

It is also possible that calls may be made for the introduction of a test for vCJD in other areas of medicine, in order to protect patients and care providers against the risk of transmission. Two such areas are surgery (especially neurosurgery) and transplantation medicine. Here too, timely consideration of the pros and cons of a test for vCJD is desirable. This is primarily a matter for the relevant professional groups and patients' organisations to consider.